Periscope

Brain

(Vol. 29, No. 114, 1906.)

1. An Address on Mendelian Heredity and Its Application to Man. W. BATESON.
2. A Contribution to the Study of Amaurotic Family Idiocy. F. J. POYNTON, J. H. PARSONS and GORDON HOLMES.
3. A Case of Orbital Encephalocele, with Unique Malformations of the Brain and Eye. J. H. PARSONS and GEORGE COATS.
4. A Study of the Various Changes which Occur in the Tissues in Acute Diphtheritic Toxemia, More Especially in Reference to Acute Cardiac Failure. LEONARD S. DUDGEON.

1. *Mendelian Heredity in Man.*—Bateson presents a brief sketch of the Mendelian hypothesis of heredity in its applications to some of the diseases of mankind, notably color blindness, congenital cataract, etc. He urges the study of hereditary diseases in the light of the Mendelian hypothesis. The paper does not permit of an abstract.

2. *Amaurotic Family Idiocy.*—The authors, from an examination of three cases of this affection, show that: (1) There is strong evidence that it is a primary disease of the nervous elements. These are affected not only throughout the whole nervous system, but also in the dorsal root ganglia and in the retina, and so universally that not a single normal cell can be found. The changes which have been described, and all the evidence that could be obtained on the mode of their development, prove that the disease is not due to a primary affection of the neuroglia, but that the proliferation of the latter has only followed the degeneration of the nervous elements. Further, there is no evidence to suggest that the changes of the nervous elements resulted from any vascular condition, there was no visible disease of the vessels and no signs of either old or recent inflammation. (2) Though they employed different methods for the demonstration and estimation of pathological changes in the different parts of the neurone, it can be asserted that the nerve cells are relatively more affected than the fibres. In many systems, as in the direct cerebellar tracts, the dorsal and ventral spinal roots, and the optic system, there may be no visible change in the fibres though the cells from which they spring are severely altered. From this fact it may be concluded that the affection is a primary cell disease, and that the alterations visible in nerve fibres are only secondary to this. (3) The condition of the nerve cells as demonstrated by the elective stains (Nissl's, Bielschowsky's), for their component parts, shows that the interfibrillar protoplasm is very much more severely affected than are the neurofibrils, and suggests that the primary change is disease of the interfibrillar protoplasm, and that to this the alterations in the neurofibrils are secondary.

Certain conclusions can also be drawn on the etiology of the disease from its morbid anatomy in conjunction with its clinical symptomatology: (1) It is not due to arrested development. If it were so, the clinical symptoms of the disease would be probably evident from birth which has not been so in any of the cases observed; and there is no reason why a mere arrest of development should produce a progressive and invariably fatal disease. There is also little anatomical evidence of mal-development, and the appearance on which Sachs lays so much emphasis can be explained by the wasting the cerebral convolutions must undergo as a result of the disease. Further, the most easily-obtained evidence of the completed development of the nervous system, myelinisation of the brain, proves that the final development of the different parts of the brain is completed at different periods in a fairly long space of time, and is not completed till a few months after birth. But the examination of these brains does not indicate greater abnormalities in the regions which develop late than in those where development is completed early in intra-uterine life; *e. g.*, the visual cortex, which is myelinated early, is quite as severely affected as the prefrontal region where the myelinated fibres appear late. If, however, the disease dates from the earlier months of extra-uterine life, the development of the fibres which myelinate late may be checked owing to deficiency of trophic influence from the diseased cells. (2) The negative results of the bacteriological examination and the entire absence of such reactions in the vascular and lymphatic systems as frequently accompany bacteriological infections suggest that the disease is not due to bacterial toxins. The clinical facts also argue against this theory; its family nature, its occurrence only in the Jewish race all militate against this view. (3) The final conclusion possible, which is supported by the microscopical investigation, is that the disease is due to some inherent biochemical property of the cells, as a result of which it undergoes certain changes which result in its degeneration, and, consecutively, in degeneration of the parts of the neurone (neurofibrils, axis-cylinders and myelin sheaths), the normal existence of which is dependent upon it. The cell changes have not the characters of a simple atrophy; in fact they seem to be due to an excessive growth of the protoplasm which later undergoes degenerative changes. This fact is not in favor of Sachs' hypothesis that the pathology of the disease can be described by the term abiotrophy (a term suggested by Gowers to represent an inherent defective vitality in the cell), or of Schaffer's suggestion that it may be explained by Edinger's "Ersatz-theorie," which assumes that elements are inherently feeble and undergo degeneration when exposed to the strain of life to which they are not normally resistant.

3. *Orbital Encephalocele.*—The authors present a detailed report of an extremely rare congenital abnormality of the eye, and an apparently unique malformation of the brain. Three congenital abnormalities of the globe were present: (1) Coloboma of the choroid, (2) Coloboma of the optic nerve entrance: the ectasia had taken place to the inner side of the intervaginal space, and the latter has not been displaced with the nerve, but has retained its normal position, (3) A central or macular coloboma. The globe abnormalities were probably due in large part to the orbital encephalocele. An isolated mass of brain substance had probably been snared off at an early period of foetal life and had interfered with the proper budding of the primary optic vesicle. It seemed probable to the writers that this alone would not explain the anomaly. Since there was no defect below, either in the nerve or in the retina, it seemed evident that a secondary optic vesicle was formed and closed in a similar manner, and that the defects in the globe were caused by the pressure of the brain

masses on the growing secondary optic vesicle. The macular coloboma remains inexplicable.

4. *Changes Due to Diphtheria Toxemia.*—Dudgeon contributes a lengthy, well-illustrated and detailed morphological study of the tissue changes in acute diphtheritic toxemia, paying particular attention to the changes thought to be responsible for acute cardiac failure. Sixteen cases served as a basis of the examinations in conjunction with extensive experimental work. The changes found in the tissues coincide with those with which pathologists are already familiar. He concludes, however, (a) That the most important lesion in the acute cases is a fatty change of the heart muscle and diaphragm, which is due to a direct action of the toxins on those tissues, (b) That similar fatty changes may be found in certain of the important viscera, especially the adrenal gland and liver, (c) That the expression "cardiac paralysis" in acute diphtheritic toxemia should be abolished and replaced by "acute cardiac failure," (d) That the changes found in the nervous system are secondary factors and not the primary cause of cardiac failure, (e) That the antitoxin, if given in sufficient quantity and within the first forty-eight hours, may prevent or considerably diminish the possibility of death from cardiac failure.

JELLIFFE.

Miscellany

FRACTURES OF BASE OF SKULL. (Annal. of Surg., December, 1906.)—Drs. L. R. G. Crandon and Louis T. Wilson analyze 530 cases of fracture of the base of the skull. The mortality was 44 per cent. Alcoholic intoxication was present in 32 per cent., of whom 38 per cent. died. Many of these developed delirium tremens, which could not be wholly differentiated from symptoms of laceration or meningitis. Forty-four per cent. were unconscious at first examination, and fully half the conscious ones were dazed or stupid. Fifty-eight per cent. of the unconscious died, against 33 per cent. of the conscious. Early delirium was usually due to alcoholism, but when late, infection was the cause. Hemorrhage from at least one cranial orifice was present in 74 per cent., the order of frequency in this series being the ear 281, nose 168, subconjunctival 53, two ears 47, mouth 44. In practically all cases of aural hemorrhage the drum was ruptured. Where pharyngeal bleeding occurs the mortality is very high. Respiration as a symptom is unimportant except in primary shock and toward a fatal termination, and the quality of the pulse is of greater significance than its rate. In 117 cases presenting stertorous breathing 106 died. A subnormal temperature seems to indicate shock or alcoholism. Nearly all those with high initial temperature died. The escape of cerebrospinal fluid was noted in only 27 cases of this series. The most striking result of the study of pupillary change was that in 142 cases without reaction 131 died. Of these 530 cases 59 were operated, with a mortality of 53 per cent. The writers conclude that only those cases should be trephined where either hemorrhage seems the most important part of the clinical picture or the course is considered one of continuous and progressive intracranial compression. Patients with non-reacting pupils, or unconscious with stertorous respiration are nearly always bleeding deep in the brain substance, and are hopeless. It is urged that in suspected basal fracture that rest in bed for three full weeks be enjoined. COWLES (New York).

CEREBROSPINAL MENINGITIS. By J. S. Billings, Jr. (Journal A. M. A., June 2).

Dr. Billings gives a history of the epidemic of cerebrospinal meningitis in New York City in 1904-05, with the methods employed to meet it. It